

CASE REPORT

Primary Intradural Hemangiopericytoma With Intramedullary Invasion

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Hemangiopericytoma (HPC) is a rare tumor of the central nervous system and is usually found intracranially. Intraspinial HPCs are very rare and mostly involve the extradural bony structures. Primary intradural HPC has only been reported in 10 cases, all of which occurred in the extramedullary region. Intramedullary invasion has never been reported. Here, we describe a case of primary intradural HPC of the thoracic spine that presented initially with paresthesia and paraplegia of both legs. Magnetic resonance imaging of the thoracic spine showed an intradural dumbbell-shaped tumor at the T10 level. The initial impression was neurogenic tumor, meningioma, or metastasis. During operation, the tumor was found to have obvious intramedullary invasion. Gross-total removal was done, and the patient's neurological function improved; there was no recurrence at the 3-year follow-up. There is no consensus as to what constitutes the optimal treatment of HPC, but most neurosurgeons will advocate gross-total resection. A comparative analysis between intradural and extradural HPCs showed a higher chance of gross-total resection for intradural HPCs, while the recurrence rates showed no difference. The role of adjuvant radiotherapy remains uncertain. Due to the high risk of recurrence and metastasis of HPCs, close follow-up for a long period is mandatory. [*J Chin Med Assoc* 2009;72(10):536–541]

Key Words: hemangiopericytoma, spinal cord, spinal neoplasms

Introduction

Hemangiopericytoma (HPC) is a tumor arising from Zimmerman's pericytes that was first described in 1942 by Stout and Murray.¹ HPC usually occurs in the subcutaneous soft tissue and skeletal system, but rarely within the central nervous system (CNS). The incidence of CNS HPC is around 1% of all CNS tumors.² Most CNS HPCs are found in the cranial cavity; they rarely occur in the spinal canal. To date, only 52 cases of intraspinal HPCs have been reported.^{3–20} Most of the intraspinal HPCs involve the extradural structures. There were only 10 intradural HPCs reported and all were extramedullary lesions.^{3,5,10,14,17} Intramedullary invasion has never been reported. HPCs are well known for their high recurrence and metastatic rates. A recent review reported a local recurrence rate of 55% and a metastatic rate of 17%.²¹ Some intraspinal HPCs are

due to recurrence or metastasis from other primary intracranial lesions,^{15,19,20} while others are primary intraspinal HPCs.

What constitutes the ideal treatment for CNS HPC remains uncertain. Most neurosurgeons suggest gross-total resection for both neural decompression and pathology diagnosis. Whether adjuvant radiotherapy would help to reduce the rate of recurrence and prolong the length of survival is unclear. Despite aggressive treatment, the 5-year recurrence rate is still around 62–65%.^{2,22}

Here, we report a case of primary intradural HPC with intramedullary invasion in the thoracic spine. It was successfully managed by gross-total resection. So far, over the course of a 3-year follow-up period, no recurrence has been found. We also review the literature on intraspinal HPCs with regard to their clinicopathological behavior, treatment, and outcome.



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Case Report

An 80-year-old, previously healthy, man was admitted to our neurosurgery department due to progressive weakness of both legs for 3 months. History-taking revealed that he had begun to suffer from numbness and a tingling sensation in both legs 6 months previously, which was followed by weakness and gait disturbance. Just prior to this hospitalization, urine incontinence occurred.

On examination, a sensory impairment below the umbilicus was found. The deep tendon reflexes of the legs were exaggerated. Paraparesis with only grade 2 muscle power of both legs was noted. An anorectal digital exam showed preserved anal tone. Ophthalmological and cranial nerve examinations were normal. Biochemical laboratory studies were unremarkable except for a mildly elevated white blood cell count ($11,700/\text{mm}^3$). An intraspinal lesion in the thoracic spine around T10 was suspected.

Magnetic resonance imaging (MRI) of the thoracic spine showed a dumbbell-shaped, posteriorly-located intradural tumor at T10 with severe cord compression. The tumor had an isointense signal on T1-weighted imaging and a hyperintense signal on T2-weighted imaging (Figure 1A). After injecting gadolinium-diethylenetriaminepenta-acetic acid (Gd-DTPA), the tumor was enhanced homogeneously (Figure 1B). Primary neurogenic tumor, meningioma or metastatic lesion was considered. A thorough survey for metastasis showed no other primary site. MRI of the rest of the spine showed an old compression fracture at the T12 vertebral body and no other significant abnormality. Brain MRI showed nothing remarkable.

The patient underwent total laminectomy from T9 to T11. After dura opening, a well-vascularized tumor was found. The tumor was mainly on the surface of the spinal cord, with intramedullary invasion (Figure 2A). The extramedullary part was removed first (Figure 2B). Next, the intramedullary part was dissected

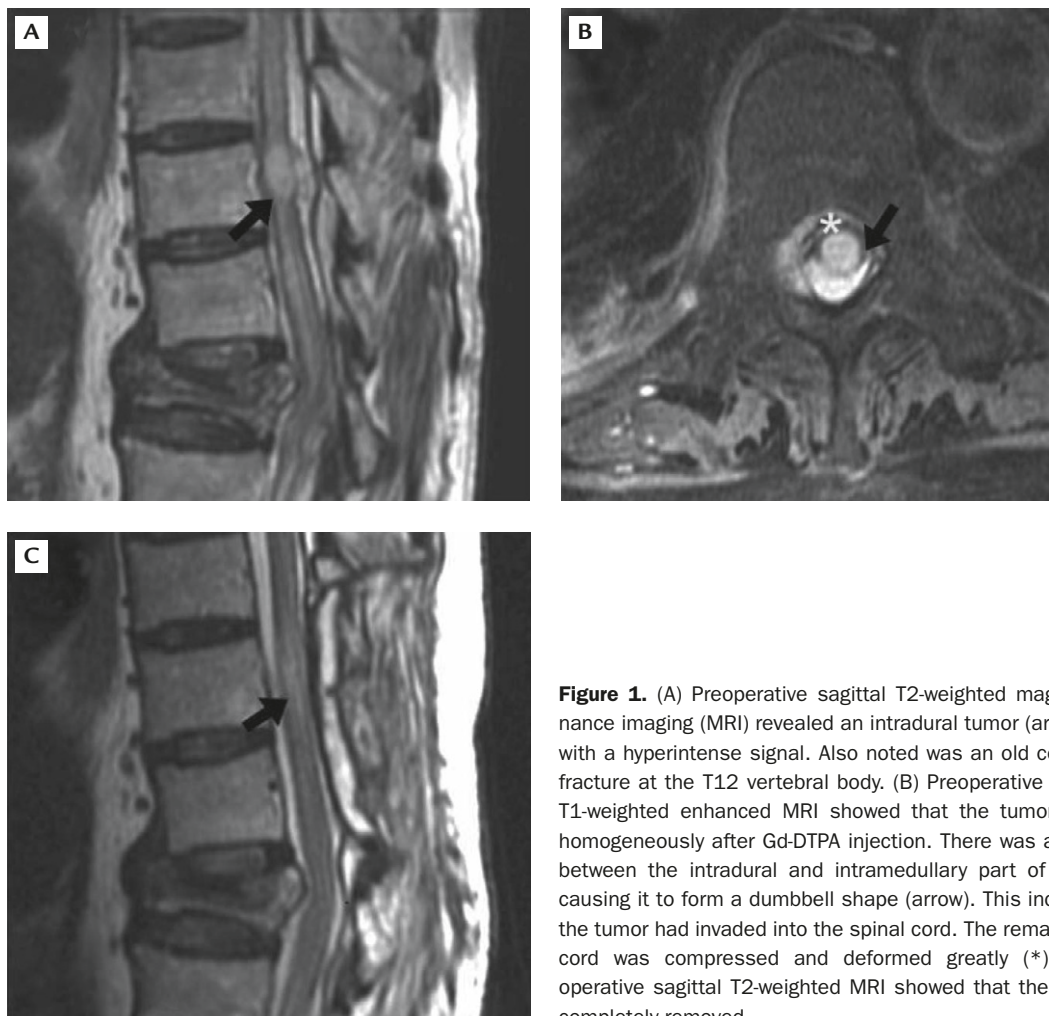


Figure 1. (A) Preoperative sagittal T2-weighted magnetic resonance imaging (MRI) revealed an intradural tumor (arrow) at T10 with a hyperintense signal. Also noted was an old compression fracture at the T12 vertebral body. (B) Preoperative axial cut of T1-weighted enhanced MRI showed that the tumor enhanced homogeneously after Gd-DTPA injection. There was a thin plane between the intradural and intramedullary part of the tumor, causing it to form a dumbbell shape (arrow). This indicated that the tumor had invaded into the spinal cord. The remaining spinal cord was compressed and deformed greatly (*). (C) Post-operative sagittal T2-weighted MRI showed that the tumor was completely removed.

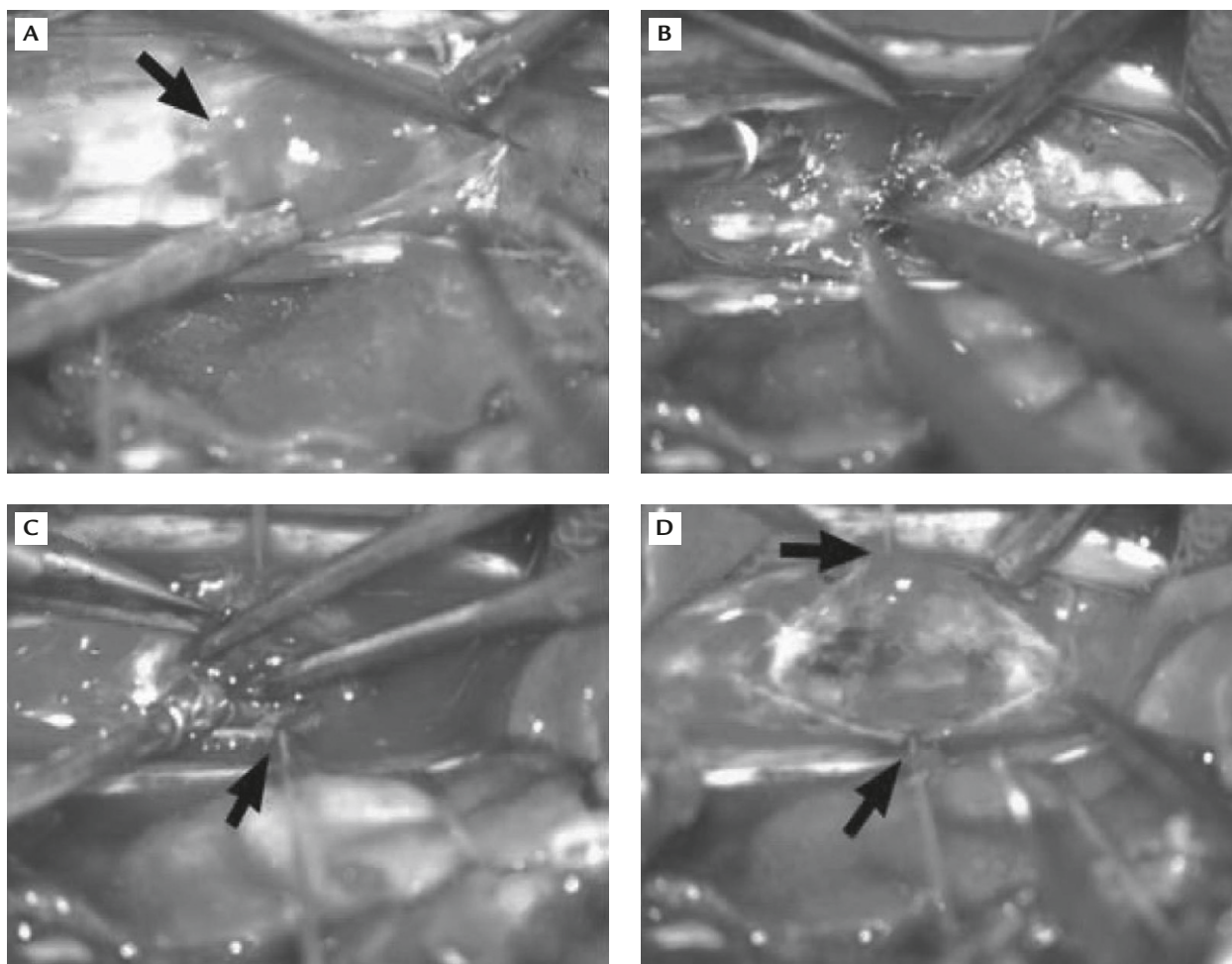


Figure 2. Intraoperative pictures of the tumor removal process. (A) After dura opening, a well-vascularized tumor mass (arrow) was found on the cord surface with intramedullary invasion. (B) The extramedullary part was removed first. (C) Next, the intramedullary part was followed into the cord tissue and removed under meticulous dissection (arrow indicates the pia opening). (D) The tumor was completely removed (arrows indicate the suture retraction of the pia mater).

into the cord tissue and removed under meticulous dissection (Figures 2C and 2D). Hemorrhaging during tumor removal was more profuse than with intradural meningioma or neurofibroma removal, but remained under control. The tumor was removed completely. Intraoperative frozen-section examination showed a well-vascularized meningioma.

Postoperatively, the patient's neurological function gradually improved. Follow-up MRI of the thoracic spine showed no residual tumor or tumor recurrence (Figure 1C). He has been followed for 3 years and continues to do well, with no evidence of disease recurrence.

The formal pathology report was as follows: grossly, the tumor was well-circumscribed, and on cut section, it was firm to rubbery with a gray-tan nodular surface. Under light microscope, the tumor was densely cellular with abundant vasculature. The vessels formed a continuous ramifying vascular network that exhibited striking variation in caliber, and some of them had

staghorn appearance (Figure 3A). The neoplastic cells proliferated between the vessels, sometimes compressing them to the point of obscuring them. The neoplastic cells were round, ovoid to slightly spindled, and had scant cytoplasm. Anaplastic features, such as hypercellularity, pleomorphism, >5 mitoses per 10 high-powered fields and necrosis, were not identified. The tumor had a rich network of reticulin fibers surrounding individual cells (Figure 3B). The tumor cells were immunoreactive for smooth muscle-specific actin (Figure 3C) and vimentin, but non-reactive for CD34 and S-100 protein. The above findings were compatible with HPC.

Discussion

HPC is a rare, highly vascularized neoplasm arising from the pericytes, the modified smooth muscle cells

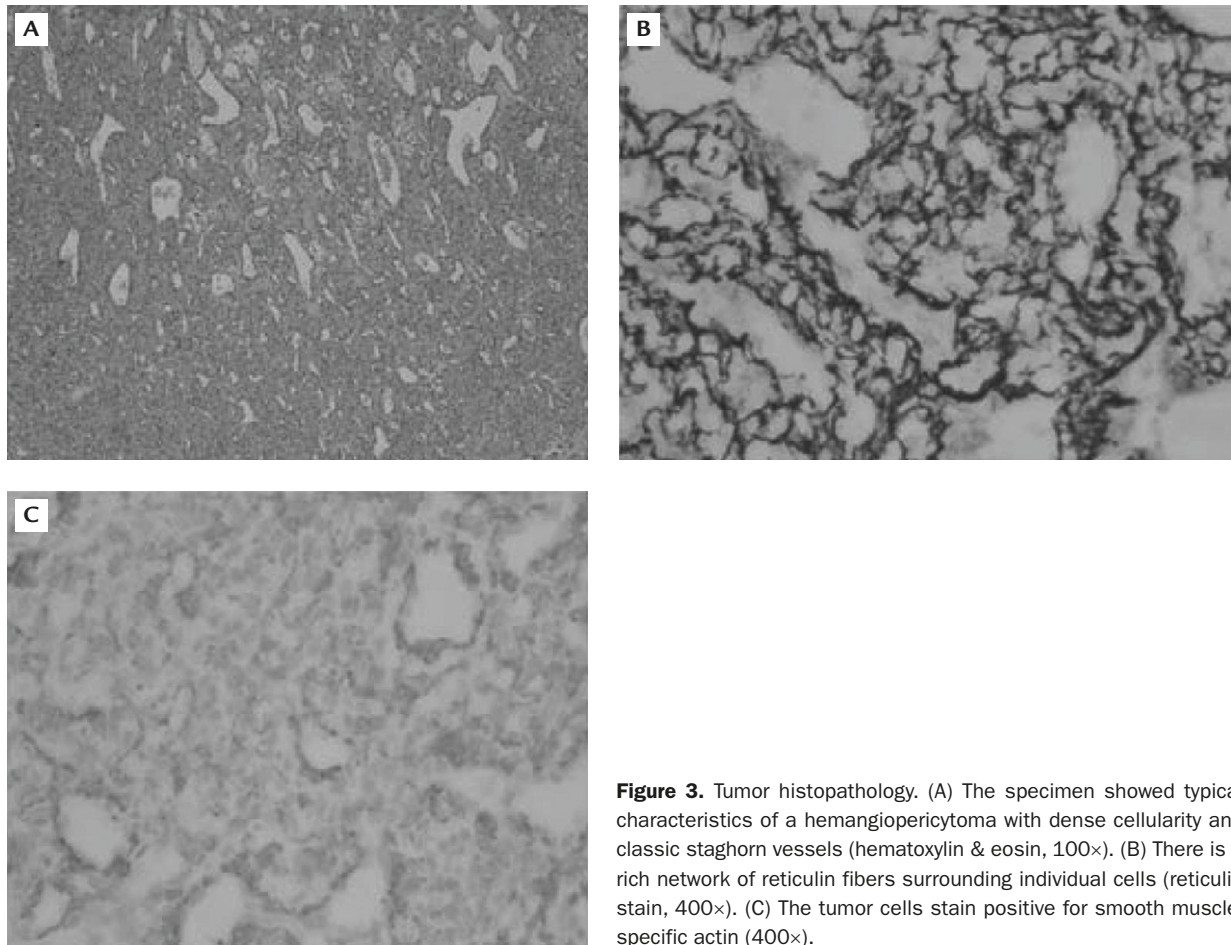


Figure 3. Tumor histopathology. (A) The specimen showed typical characteristics of a hemangiopericytoma with dense cellularity and classic staghorn vessels (hematoxylin & eosin, 100 \times). (B) There is a rich network of reticulin fibers surrounding individual cells (reticulin stain, 400 \times). (C) The tumor cells stain positive for smooth muscle-specific actin (400 \times).

that normally surround reticular sheath capillaries.¹ HPC usually occurs in the subcutaneous soft tissue and skeletal system, and rarely within the CNS. A number of series of meningeal HPCs have already been reported.^{2,10,12,21,23,24} For intraspinal HPCs, due to their rarity, only several case reports and small case series were found. A literature review showed that there were 52 intraspinal HPCs reported, including 41 extradural HPCs and 11 intradural HPCs (including the present case).^{3–20} Eighteen were in the cervical spine, 22 in the thoracic spine (including the present case), 5 in the lumbar spine, 1 in the sacrum, and 6 were unknown.^{3–20} Most of the reported intraspinal HPCs were included in the meningeal HPC series without any detailed descriptions of their clinical behavior, treatment and outcome.^{2,12,23,24} Guthrie et al in 1989 reported a series of 44 meningeal HPCs, with only 2 in the spine.² A series reported in 2003 by Ecker et al included 38 HPCs, with 3 found in the spine.²⁴ In 1958, Schirger et al were the first to describe this neoplasm of the spine.¹⁵ Since then, 51 additional cases have been reported.^{3–14,16–20} Most of the intraspinal HPCs occurred in the extradural region

such as the vertebral body, lamina, pedicle or epidural space. In 1961, Kruse¹⁰ was the first to report an intradural spinal HPC. Since then, 9 additional cases have been reported.^{3,5,14,17} All of them were found in the intradural extramedullary region.

The MRI characteristics of intraspinal and intracranial HPCs are the same. They usually have an isointense signal on T1-weighted imaging and hyperintense signal on T2-weighted imaging. After Gd-DTPA injection, HPCs enhance homogeneously.^{2,3,17} When an extradural HPC grows and expands, it sometimes erodes the adjacent bony structures such as the lamina, pedicle, neural foramen and even paraspinal muscle. The expansion of extradural HPCs through the neural foramen will form the characteristic dumbbell-shape appearance on MRI.¹⁷ Since HPC is a tumor that arises from the pericytes of capillaries, it is possible that HPC may follow the cord surface vessels to invade cord tissue, as was observed in the present case. This is different from intradural meningioma or neurofibroma, which occur purely in the extramedullary space and have a good surgical “plane” during dissection. The preoperative MRI of the thoracic spine in our patient

showed a dumbbell-shaped tumor with suspected intramedullary invasion. This is different from the known expansion type which is usually observed in other extradural HPCs because in the present case, the dumbbell shape was observed purely in the *intradural* region. To the best of our knowledge, there are no previous reports with this type of finding. We carefully inspected the relation of the tumor and spinal cord and then dissected inside the cord for total removal. In previous articles, most neurosurgeons recommend *en bloc* resection to reduce intraoperative blood loss and recurrence rate.^{2,3,21} But when tumor with severe neural compression is encountered, central debulking and piecemeal removal may be needed to reduce neural damage, which is what we did in our case. Significant intraoperative blood loss used to be a major problem.^{2,5,13,22,24} But by utilizing modern microsurgical techniques, improved preoperative imaging surveys and better neurointensive care, perioperative mortality and morbidity rates have been reduced a lot.

A further analysis of the 52 intraspinal HPCs showed that there were 25 extradural HPCs and 9 intradural HPCs with follow-up data. The overall recurrence rate was 53%. There were 14 (56%) recurrences in the extradural HPC group and 4 (44%) in the intradural HPC group. Statistical analysis using Fisher's exact test showed no significant difference ($p=0.42$). Further analysis between extradural and intradural HPCs showed that intradural HPCs harbor a higher chance of gross-total removal. Gross-total removal was achieved in 9 cases (including the present case) among all 11 (82%) intradural HPCs, which was significantly higher than the 15 of 41 (38%) extradural HPCs ($p<0.05$). The lower rate of gross-total resection of extradural HPCs may be due to their firm adherence to the dura mater, irregular invasion of the bone, and rich vascularization. Preoperative embolization may help in bleeding control and better removal.^{5,12,13} It should be remembered, however, that if severe compression already exists, embolization may lead to clinical deterioration, probably caused by tumor swelling, and emergent surgery may be needed. Also, after extensive removal of extradural HPC, some type of fusion and fixation device may be needed to reconstruct the destroyed bony structure.¹⁶ All of these issues make extradural HPC removal difficult. The surgical strategy for intradural HPC removal is much simpler as long as the HPC can be freed from neighboring nerve tissue. Guthrie et al² reported that total tumor resection favorably affected recurrence and survival, although this did not reach statistical significance. In a recent series reported by

Ecker et al,²⁴ gross-total resection did not provide any significant survival benefit. Our analysis showed that gross-total resection had no benefit in reducing recurrence rate. A possible reason for this may be that HPC is a highly malignant tumor that is prone to recurrence and metastasis. Standard surgical eradication may fail to control disease progression. Furthermore, recurrence and metastasis may occur very late after the first operation.^{15,19,20,25} A long follow-up period is thus mandatory.

Radiotherapy was once advocated to reduce recurrence rate and improve length of survival for CNS HPCs.^{2,12,22,26-28} Guthrie et al found that adjuvant radiation therapy extended the average time before first recurrence and extended the survival period,² but Ecker et al found no survival benefit from radiotherapy.²⁴ We analyzed 16 patients with intraspinal HPCs who received adjuvant radiotherapy;^{7-10,13,15,17,19,20} 9 had follow-up data, 5 of whom suffered from recurrence (56%). There was no significant difference from those who had not received radiotherapy (52%, $p=0.58$). The role of radiotherapy is still uncertain. Recently, stereotactic radiosurgery emerged as a useful tool for intracranial HPCs. In 1993, Coffey et al²⁹ reported the first series of patients in whom radiosurgery was used to treat recurrent HPCs; an 82% tumor control rate was noted. Payne et al³⁰ reported an 87% tumor control rate during a mean follow-up of 2.5 years, and Ecker et al²⁴ reported a 93% control rate during a mean follow-up of 3.8 years. Radiosurgery may be a good alternative to repeated surgery in residual or recurrent cases.

In conclusion, we have reported a case of primary intradural HPC with intramedullary invasion. Gross-total removal was achieved, and no recurrence has been noted up to the present time. A literature review showed that intradural HPCs have a higher chance of total resection. There is no difference in recurrence rate between extradural and intradural HPCs. Radiotherapy does not appear to help in reducing recurrence rates. The rarity of this tumor in the spine, specifically in the intradural region, makes further studies necessary. The ideal treatment has yet to be determined, but should be individualized for each specific case.

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